

## Case Report

Takayasu's Arteritis in a Patient with Sydenham's Chorea:  
is There an Association?Thiago Cardoso Vale<sup>1</sup>, Ricardo Oliveira Horta Maciel<sup>1</sup>, Débora Maia<sup>1</sup>, Rogério Beato<sup>1</sup> & Francisco Cardoso<sup>1\*</sup><sup>1</sup> Movement Disorders Clinic, Faculty of Medicine, Universidade Federal de Minas Gerais, Belo Horizonte, Brazil

## Abstract

**Background:** Takayasu's arteritis (TA) has been associated with many conditions. Herein described is a case of TA in a patient with rheumatic fever complicated with Sydenham's chorea.

**Case Report:** A 17-year-old female presented at age 6 with rheumatic fever followed by chorea a month later. At the age of 16, she developed a blood pressure discrepancy between the arms and faint pulses. Computed tomography angiography revealed diffuse aortic involvement and narrowing of the arteries.

**Discussion:** The presence of rheumatic fever and Sydenham's chorea in TA raises the possibility of an immunological basis for the pathogenesis of the disease.

**Keywords:** Chorea, Sydenham's chorea, Takayasu's arteritis, rheumatology, rheumatic fever, hyperkinetic movement disorders

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## Introduction

Takayasu's arteritis (TA) is a rare chronic large-vessel vasculitis of unknown etiology. First reported by the Japanese ophthalmologist Takayasu in 1908, TA has an age of onset between the second and fifth decades of life, and females are more commonly affected than males, in an estimated ratio of 6.6:1.<sup>1–3</sup> Since the original description, the disease has been associated with various conditions, including rheumatoid arthritis and other collagen vascular diseases, syphilis, tuberculosis, rheumatic fever and other streptococcal infections, mitral stenosis, and rheumatic heart disease.<sup>3–5</sup> Its association with rheumatoid arthritis and lupus erythematosus disease raises the possibility of a shared primary or secondary autoimmune mechanism contributing to the pathogenesis of the disease.<sup>5</sup>

Rheumatic fever (RF) is an autoimmune disease related to previous throat infection with group A  $\beta$ -hemolytic *Streptococcus*, but rarely associated with TA.<sup>1</sup> One of the late complications of RF is Sydenham's chorea (SC), a hyperkinetic movement disorder, which occurs as a result of dysfunction of basal ganglia structures mediated by cross-reactive antibodies to *Streptococcus*.<sup>6</sup>

We aim to describe a case of TA in a patient with a previous history of RF complicated with SC. The presence of RF and SC in TA raises the possibility of a common immunological basis for the pathogenesis of both diseases.

## Case report

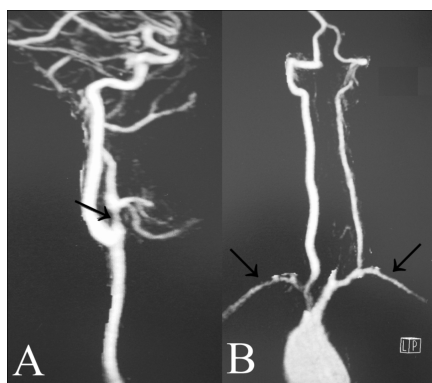
A 17-year-old female was admitted to our Movement Disorders Clinic at age 6, when she presented acutely with fever, weight loss, tachycardia, and bilateral knee and ankle arthritis. Cardiovascular examination was unremarkable. A month later, she developed involuntary movements of her face, trunk and limbs, particularly on the right side. She had difficulties in writing and holding objects in her hands. Her speech was moderately impaired and there were evident behavioral abnormalities, such as apathy and emotional incontinence. She was diagnosed as having RF complicated by SC, according to the modified Jones criteria,<sup>7</sup> after having other causes of chorea ruled out (drug-induced, metabolic and toxic encephalopathies, other infectious choreas, systemic lupus erythematosus, post-vaccinal encephalitis, paraneoplastic choreas, and genetic such as benign hereditary chorea, ataxia–telangiectasia, and Huntington's disease).

She received haloperidol (5 mg/day) with improvement after 1 month and complete resolution in 3 months. She was also given penicillin G benzathine every 21 days as prophylaxis for new episodes of *Streptococcus* infection.

At the age of 14, she developed another bout of mild choreic movements, which resolved in 3 months after a short course of haloperidol (2 mg/day). At the time, a clinical cardiologic examination was unrevealing, but echocardiography disclosed a thickened mitral valve causing mild mitral regurgitation and mild valve prolapse. At the age of 16, on a regular follow-up examination, she was found to have bilaterally faint radial and brachial pulses and a 20 mmHg discrepancy in the systolic blood pressure between the arms. Cervical ultrasonography disclosed diffuse thickened subclavian arteries. Computed tomography angiography showed diffuse aortic involvement with mild stenosis of the mesenteric, common hepatic, and right renal arteries. Both subclavian arteries and the left external carotid artery also had narrowing and contour abnormalities. In addition, there was a moderate stenosis of the proximal portion of the right external carotid artery (Figure 1). Brain magnetic resonance imaging was normal. Antinuclear antibodies, antineutrophil cytoplasmic antibodies, lupus anticoagulant, and anticardiolipin antibodies were negative. The erythrocyte sedimentation rate was 69 mm/hour (normal value <13 mm/hour). She fulfilled the 1990 American College of Rheumatology criteria for TA and was classified as having type III angiographic features according to the Takayasu Conference Classification.<sup>2,9</sup> She was treated with methotrexate (7.5 mg four times a week) combined with prednisone (60 mg/day).

### Discussion

TA is a systemic necrotizing vasculitis that results in post-inflammatory stenosis or occlusions of large vessels, predominantly of the aorta and its branches. Pathologically, the disease manifests with granulomatous inflammation of the vessel adventitia and the outer part of the media, with infiltration of lymphocytes, plasma cells, histiocytes, and multinucleated giant cells.<sup>1,8,9</sup>



**Figure 1. Computed Tomography Angiography of the Neck.** (A) Bifurcation of the right common carotid artery showing area of stenosis (arrow) in the proximal external carotid artery. (B) Narrowing and contour abnormalities (arrows) in both subclavian arteries.

Diagnosis of TA rests on clinical suspicion in any young patient, particularly women, with clinical manifestations of vascular ischemia and the presence of bruits, decreased or absent pulses, ischemic ulcers, or a combination of these findings. In children, the usual presenting features are hypertension, heart failure, or a neurological symptom.<sup>8</sup> Claudication, bruit, or a faint pulse in an asymptomatic child are uncommon presentations,<sup>8</sup> as in our case. TA does have a prolonged, relapsing, and remitting course; in cases of unsuccessful treatment with corticosteroids, treatment with cytotoxic agents, such as cyclophosphamide or methotrexate, should be considered.<sup>1</sup>

The co-occurrence of RF and TA has infrequently been described. Tejada et al.<sup>10</sup> reported two cases of rheumatic heart disease within a series of 125 patients diagnosed with TA. Autopsy studies from India found pathological evidence of rheumatic myocarditis in one patient out of 10.<sup>11</sup> No SC cases were present in any of these reports.

Currently, the evidence suggests that the pathogenesis of SC is related to circulating cross-reactive antibodies, formed by streptococcal infection in genetically predisposed subjects, leading to disruption of the basal ganglia function. Several studies have demonstrated the presence of such circulating antibodies in 50–90% of patients with SC.<sup>6</sup> The precise factors in TA that are responsible for the vascular damage are unknown. The presence of hypergammaglobulinemia, circulating antibodies against aorta and arteries, circulating immune complexes, and a favorable response to steroids suggests the pathogenic role of autoimmunity.<sup>8</sup> One can speculate that in patients with RF and TA, serum antibodies could recognize cardiac, brain and vessel antigens causing both conditions.

Although it remains to be seen whether our case represents an etiological relation or just a coincidental association, the presence of RF and SC in a patient with TA raises the possibility of a common immunological basis for the pathogenesis of both diseases. To our knowledge, TA in the context of previous RF complicated with SC has never been reported before.

### References

1. Brunner J, Feldman BM, Tyrrell PN, et al. Takayasu arteritis in children and adolescents. *Rheumatology (Oxford)* 2010;49:1806–1814, <http://dx.doi.org/10.1093/rheumatology/keq167>.
2. Arend WP, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. *Arthritis Rheum* 1990;33:1129–1134, <http://dx.doi.org/10.1002/art.1780330811>.
3. Doi YL, Seo H, Hamashige N, et al. Takayasu's arteritis and mitral stenosis. *Clin Cardiol* 1988;11:123–125, <http://dx.doi.org/10.1002/clc.4960110212>.
4. Lupi-Herrera E, Sanchez-Torres G, Marcushamer J, et al. Takayasu's arteritis. Clinical study of 107 cases. *Am Heart J* 1977;93:94–103, [http://dx.doi.org/10.1016/S0002-8703\(77\)80178-6](http://dx.doi.org/10.1016/S0002-8703(77)80178-6).
5. Nakao K, Ikeda M, Kimata S, et al. Takayasu's arteritis. Clinical report of eighty-four cases and immunological studies of seven cases. *Circulation* 1967;35:1141–1155, <http://dx.doi.org/10.1161/01.CIR.35.6.1141>.
6. Cardoso F. Sydenham's chorea. *Handb Clin Neurol* 2011;100:221–229, <http://dx.doi.org/10.1016/B978-0-444-52014-2.00014-8>.

7. Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. *JAMA* 1992;268:2069–2073, <http://dx.doi.org/10.1001/jama.1992.03490150121036>.
8. Gulati A, Bagga A. Large vessel vasculitis. *Pediatr Nephrol* 2010;25:1037–1048, <http://dx.doi.org/10.1007/s00467-009-1312-9>.
9. Minagar A, Fowler M, Harris MK, et al. Neurologic presentations of systemic vasculitides. *Neurol Clin* 2010;28:171–184, <http://dx.doi.org/10.1016/j.ncl.2009.09.015>.
10. Tejada O, Torres S, Alfaro K. Cardiac damage in Takayasu's arteritis. Study in 125 patients. *Arch Inst Cardiol Mex* 1983;53:441–447.
11. Sharma BK, Jain S, Radotra BD. An autopsy study of Takayasu arteritis in India. *Int J Cardiol.* 1998;66:S85–90, [http://dx.doi.org/10.1016/S0167-5273\(98\)00155-7](http://dx.doi.org/10.1016/S0167-5273(98)00155-7).

